

MPDU1 siRNA (m): sc-149527

BACKGROUND

MPDU1 (mannose-P-dolichol utilization defect 1 protein), also designated suppressor of Lec15 and Lec35 glycosylation mutation or SL15, mediates the transfer of glucose and mannose residues from Glc-P-Dol and Man-P-Dol to oligosaccharides. Defects in the MPDU1 gene result in a type I congenital disorder of glycosylation CDG-If. Patients with CDG-If make incomplete lipid-linked oligosaccharides (LLO) and present with severe psychomotor retardation, seizures, failure to thrive, dry skin and scaling with erythroderma and impaired vision. Overexpression of GlcNAc-1-P transferase has been shown to impair the function of MPDU1, suggesting a form of pseudo-CDG-If.

REFERENCES

1. Ware, F.E. and Lehrman, M.A. 1996. Expression cloning of a novel suppressor of the Lec15 and Lec35 glycosylation mutations of Chinese hamster ovary cells. *J. Biol. Chem.* 271: 13935-13938.
2. Kranz, C., Denecke, J., Lehrman, M.A., Ray, S., Kienz, P., Kreissel, G., Sagi, D., Peter-Katalinic, J., Freeze, H.H., Schmid, T., Jackowski-Dohrmann, S., Harms, E. and Marquardt, T. 2001. A mutation in the human MPDU1 gene causes congenital disorder of glycosylation type If (CDG-If). *J. Clin. Invest.* 108: 1613-1619.
3. Schenk, B., Imbach, T., Frank, C.G., Grubenmann, C.E., Raymond, G.V., Hurvitz, H., Korn-Lubetzki, I., Revel-Vik, S., Raas-Rotschild, A., Luder, A.S., Jaeken, J., Berger, E.G., Matthijs, G., Hennen, T. and Aepli, M. 2001. MPDU1 mutations underlie a novel human congenital disorder of glycosylation, designated type If. *J. Clin. Invest.* 108: 1687-1695.
4. Dupre, T., Lavieue, G., Moore, S. and Seta, N. 2004. Inherited disorders of protein glycosylation. *Med. Sci.* 20: 331-338.
5. Eklund, E.A., Merbouh, N., Ichikawa, M., Nishikawa, A., Clima, J.M., Dorman, J.A., Norberg, T. and Freeze, H.H. 2005. Hydrophobic Man-1-P derivatives correct abnormal glycosylation in Type I congenital disorder of glycosylation fibroblasts. *Glycobiology* 15: 1084-1093.
6. Freeze, H.H. 2007. Congenital disorders of glycosylation: CDG-I, CDG-II, and beyond. *Curr. Mol. Med.* 7: 389-396.
7. Gao, N., Shang, J. and Lehrman, M.A. 2008. Unexpected basis for impaired Glc3Man9GlcNAc2-P-P-dolichol biosynthesis by elevated expression of GlcNAc-1-P transferase. *Glycobiology* 18: 125-134.

CHROMOSOMAL LOCATION

Genetic locus: Mpdu1 (mouse) mapping to 11 B3.

PRODUCT

MPDU1 siRNA (m) is a target-specific 19-25 nt siRNA designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see MPDU1 shRNA Plasmid (m): sc-149527-SH and MPDU1 shRNA (m) Lentiviral Particles: sc-149527-V as alternate gene silencing products.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNases and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

MPDU1 siRNA (m) is recommended for the inhibition of MPDU1 expression in mouse cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 μ M in 66 μ l. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor MPDU1 gene expression knockdown using RT-PCR Primer: MPDU1 (m)-PR: sc-149527-PR (20 μ l). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.